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## Glial heterotopia of the oral cavity



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## ABSTRACT

We report an unusual case of a glial heterotopia arising from the oral cavity of an African neonate. The patient presented with an external pedunculated oral mass which was connected to the anterior hard palate by a firm, rubbery stalk of mucosal tissue. While the mass appeared painless, it interfered with the infant's feeding and was disturbing to the parents. After a computed tomography scan excluded an intracranial connection, the mass was excised at its base and sent for biopsy. Histopathology examination confirmed glial heterotopia. Glial heterotopias should be included in the differential diagnosis of congenital masses in the oral region.

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Congenital oral lesions occur infrequently in newborns. The most commonly encountered oral lesions are keratin cysts, such as Epstein pearls and Bohn nodules; however, the differential diagnosis is broad and includes disorders such as polyps, dermoids, teratomas, sarcomas or encephaloceles. This report describes the rare case of a female infant with a pedunculated mass originating from her anterior hard palate that proved to be a glial heterotopia.

## 1. Case report

A two-week old female presented to Tamale Teaching Hospital, Tamale, Ghana with a painless mass extending outwards from the midline of the anterior hard palate (Fig. 1, Panels A & B). Her physical examination was unremarkable except for hypertelorism. The parents reported that the mass was increasing in size and interfering with breastfeeding. Computerized tomography of the head did not show that the mass had any clear continuity with intracranial structures. After consenting the family regarding the

risks and benefits of surgery, the mass was suture ligated at its base and then excised without any noticeable CSF extravasation. The patient was discharged home on the second post-operative day tolerating feedings. She has since been lost to follow up.

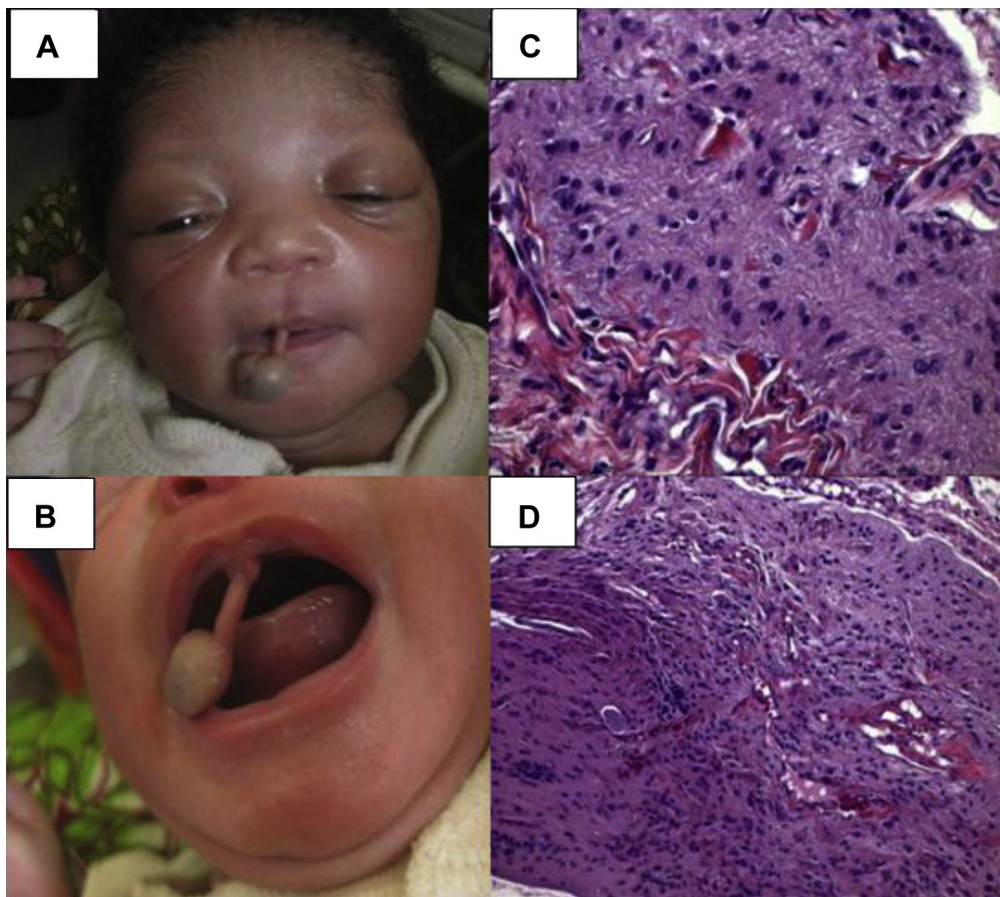
Histopathological examination revealed a polypoid mass with a surface that consisted of keratinized stratified squamous epithelium (Fig. 1, Panels C & D). The central portion of the mass contained a small island of glial tissue with astrocytes and rare neurons, consistent with brain tissue. There were also some irregular strands of connective tissue with epithelial appearing cells adjacent to the glial tissue suggestive of meninges. The stalk had prominent arteries and veins with adjacent myxoid changes. There was no evidence of cytologic atypia or malignancy. Immunohistochemical staining was positive for glial fibrillary acidic protein (GFAP) and negative for epithelial membrane antigen, consistent with the diagnosis of a glial heterotopia.

## 2. Discussion

Heterotopia refers to tissue that occurs in an abnormal location. Glial heterotopias are collections of normal glial tissue that are found in an abnormal location without intracranial connections. Glial heterotopias are best categorized as choristomas, a general

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**Fig. 1.** Mass (A & B) extending from the anterior hard palate via a midline stalk of firm, rubbery mucosal tissue. (C) Glial tissue with astrocytes and neutrophils in the background (200× H&E). (D) Glial tissue with neuron cytoplasm from the central area of polyp (100× H&E).

term that describes collections of normal tissue found in an ectopic location [1]. Glial heterotopias are embryologically related to encephaloceles, but are distinguishable because they do not connect to the central nervous system [2].

Glial heterotopias are typically midline structures and most commonly occur in the nasal cavity but have also been described involving the soft palate [3], tongue [4], hard palate [5], nasopharynx [6], upper lip [7], orbit [1], head and neck [8] and lungs [9]. Sixty percent of gliomas are extranasal, thirty percent are intranasal, and ten percent are both [10]. Oral glial heterotopic lesions usually affect the neonate's ability to feed and these lesions are extremely rare.

Oral glial heterotopias are rare benign entities and, without gender or ethnic predispositions, are generally reported in the palatopharyngeal complex area of infants followed by the tongue [2]. There are no known syndromic predispositions or identified etiologic factors. The pathogenesis is thought to result from herniation or protrusion of neural tissues through an arrested closure in the osseous cranium tissue that becomes isolated from the brain in later development [11].

Glial heterotopias can be confused with other congenital oral masses such as polyps, cysts, dermoids or encephaloceles. Thus, in addition to physical exam, imaging during the preoperative workup is important to determine whether the mass has any intracranial connections because meningitis or CSF leaks are serious complications if not recognized [1]. The complications of oral glial heterotopias depend on their site of occurrence and size of the lesion and can include airway obstruction, feeding difficulties and cosmetic

distortions [8]. Computed tomography scans can determine the location and extent of the mass and rule out any bony erosion or skull base defects. If the CT scan excludes any intracranial extent, resection via excisional biopsy should be performed. If the scan shows a skull base defect, an MRI can be performed to rule out any intracranial communication [12].

The treatment of choice for oral heterotopias is complete surgical excision without compromising function or injuring adjacent structures [5]. Early surgical intervention is favored as it decreases the chance of distortion of facial bones or abnormal development of swallow function and pharyngeal coordination [13]. If continuity does exist with the central nervous system, a craniotomy is needed to close the defect [14]. The rare case of recurrence is typically due to incomplete resection.

A diagnosis of a glial heterotopia is confirmed histologically by the presence of neuroglial tissue intermixed with neuron cells, with or without choroids plexus and clefts lined by ependymal-type epithelium, which is powerfully positive for GFAP and S-100 proteins while weakly positive for nestin and vimentin [15].

### 3. Conclusion

Oral glial heterotopias should be included in the differential diagnosis of congenital oral malformations. Early diagnosis and treatment can reduce complications and minimizes the chance of facial bone distortion. Isolated oral heterotopias have an excellent prognosis with surgical resection usually curative.

**Conflict of interest statement**

No authors have any disclosures.

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